

## REVIEW ARTICLE

# Vascular Problems of the Pelvis

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Vascular tumors of the pelvis are a rare, diverse group of neoplasms. These benign or malignant tumors can arise from the endothelium, smooth muscle cells, or pericytes of the arterial venous or lymphatic walls. They are rarely diagnosed by physical examination but more commonly seen with imaging studies such as computed tomography, magnetic resonance imaging, or angiography. Benign and malignant tumors can be differentiated pathologically by the two major anatomic characteristics of vascular channel formation and the regularity of endothelial cell proliferation. This review will focus on the clinical presentation, diagnosis, treatment, outcomes, and surgical approaches to benign and malignant vascular tumors of the pelvis.

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**KEY WORDS:** vascular tumors; pelvis; surgical approaches

## INTRODUCTION

Vascular tumors are a diverse group of neoplasms that arise from the endothelium, smooth muscle cells, and pericytes of the arterial, venous, or lymphatic walls. This review focuses on vascular tumors arising in the pelvis. Vascular tumors of the pelvis are rarely detected on physical examination. They are more commonly diagnosed when symptoms prompt diagnostic imaging procedures such as computerized tomography (CT), magnetic resonance imaging (MRI), ultrasonography, radiography, arteriography, or venography. The differential diagnosis in a patient with a pelvic mass is quite extensive and includes some rare vascular tumors. Vascular tumors of the pelvis can be both benign and malignant (Table I). The pathological delineation between a benign and malignant vascular tumor of the pelvis can be difficult and is based on two major anatomic characteristics of the tumor: (1) the formation of vascular channels and (2) the regularity of endothelial cell proliferation [1].

Vascular tumors of the pelvis often cause significant morbidity and mortality, whether benign or malignant. Benign tumors of the pelvis can cause venous compression, deep vein thrombosis, leg swelling, testicular swelling, and intestinal obstruction. Malignant neoplasms cause significant morbidity, with local tumor invasion

and death due to metastases. Death can also occur in patients with benign tumors of the pelvis secondary to ureteral or intestinal obstruction and deep vein thrombosis from pulmonary embolism. A clear understanding of the clinical presentation, diagnostic options, and treatment for benign and malignant vascular tumors is extremely important in minimizing morbidity and mortality. This review describes the diagnosis, treatment, and outcome of patients with benign and malignant vascular tumors of the pelvis.

## BENIGN TUMORS

Vascular tumors of the pelvis are considered benign based on the absence of invasion and metastases and the presence of well-formed vessels and regular endothelial cells. Benign vascular tumors are of multiple cell line origins and include hemangiomas, endolymphatic stromal myosis, lymphangioleiomyomatosis, angiomyxomas, and hamartomas. Arterial and venous aneurysms, although not neoplasms, and arteriovenous malforma-

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TABLE I. Vascular Tumors of the Pelvis

| Benign aneurysms                  | Malignant                   |
|-----------------------------------|-----------------------------|
| Arterial                          | Hemangiopericytoma          |
| Iliac artery                      | Leiomyosarcoma              |
| Sciatic artery                    | Angiosarcoma                |
| Venous                            | Lymphangiosarcoma           |
| Iliac                             | Intimal sarcoma             |
| Varix                             |                             |
| Klippel-Trenaunay syndrome        | Hemangioendothelioma        |
| Iliac vein compression/atresia    | Epithelioid                 |
| Arterial venous malformations     | Retiform                    |
| Hemangiomas                       | Papillary                   |
| Capillary                         | Kaposi-like                 |
| Cavernous                         | Intravenous tumors          |
| Deep soft tissue                  | Endometrial stromal sarcoma |
| Lymphocyst                        | Leiomyomatosis              |
| Angiomyxoma                       | Renal cell carcinoma        |
| Hamartomas                        | Myxoma/sarcoma              |
| Glomus tumor                      | Lymphangiioleiomyomatosis   |
| Spindle cell hemangioendothelioma | Kaposi sarcoma              |

tions and lymphocysts are important vascular tumors of the pelvis that are discussed in the sections that follow.

### Arterial Aneurysms

Most aneurysms of the large arteries of the pelvis involve both the aorta and iliac arteries. Abdominal aortic aneurysms are calcified in nearly 80% of patients, but iliac artery aneurysms are calcified in only approximately 35% of patients. Isolated iliac artery aneurysms occur in less than 2% of patients [2,3], typically involving the common or internal iliac artery and rarely involving the external iliac artery (Fig. 1A). A pulsatile abdominal mass may not be detected on abdominal examination because of its location in the pelvis; however, it may be detected by a palpable, pulsatile mass on rectal examination in the buttock area [2–4]. In addition, thrombosis of the aneurysms can make differential diagnosis difficult, especially for sciatic artery aneurysms, if the congenital abnormality is not known. Sciatic artery aneurysms more commonly embolize to the extremity, thrombose, or less frequently, occur with rupture.

Patients with isolated iliac artery aneurysms of the pelvis may have symptoms caused by compression of adjacent structures or rupture. Symptomatic patients may have urinary symptoms such as urgency and frequency, intestinal symptoms such as constipation, or extremity swelling caused by vein compression (Fig. 1B). Erosion of iliac artery aneurysms into the ureter as well as the rectosigmoid colon has also been reported in patients with massive hematuria or lower gastrointestinal bleeding. Patients with a ruptured iliac artery aneurysm often have back, hip, buttock, or testicular pain as well as the presence of hematoma of the flank or scrotum. Isolated

aneurysms of the iliac artery may also be asymptomatic. The diagnostic test of choice in patients with suspected iliac artery aneurysms is a CT scan of the abdomen and pelvis.

Symptomatic and asymptomatic aneurysms that are 4 cm or greater in diameter should be electively repaired. Asymptomatic iliac artery aneurysms that are greater than 4 cm in diameter should be repaired due to the difficulty in diagnosis, access to the aneurysms, and the risk of rupture [5]. Endoaneurysmorrhaphy is the preferred treatment, as with abdominal aortic aneurysms. For patients with isolated internal iliac artery aneurysms, proximal ligation alone is not adequate, especially in patients with symptoms caused by the mass effect of the aneurysm on the adjacent organs. Internal iliac artery aneurysms should be ligated proximally and then opened with evacuation of the clot and ligation of all branches from within the aneurysm. With bilateral iliac artery aneurysms, care to preserve flow to at least one of the internal iliac arteries is essential as well as to the inferior mesenteric artery in order to prevent subsequent pelvic necrosis or paraplegia [6,7]. Sciatic artery aneurysms should be repaired with ligation of the aneurysm proximally and distally through a gluteal incision and subsequent revascularization via femoropopliteal bypass. The operative mortality is less than 10% for an elective repair of the iliac artery aneurysm and greater than 60% for ruptured artery aneurysms.

### Venous Aneurysms

Primary venous aneurysms are rare congenital abnormalities, but occasionally do occur in the pelvis. Dilatation of the iliac vein or its branches can appear as a pelvic mass and must be considered in the differential diagnosis. Figure 2 represents a patient who was asymptomatic with an incidental finding of a pelvic mass, which on venography, color duplex ultrasonography, and CT scan was an iliac vein aneurysm. Venous aneurysms can also occur at multiple other locations [8]. Venous aneurysms have been reported in association with deep vein thrombosis as well as pulmonary embolus. No treatment is indicated or recommended in patients who are asymptomatic. Dilatation of the iliac veins in the pelvis or its branches can also occur in association with cirrhosis and venous hypertension [9], iliac pain, hypoplasia, and Klippel-Trenaunay syndrome, in which there is hypoplasia of the external iliac vein, with marked distention and collateralization of the internal iliac vein and common iliac vein [10].

### Arteriovenous Malformations

Arteriovenous malformations of the pelvis can be congenital or secondary to previous interventional procedures or trauma. Congenital vascular malformations may range from the capillary to large vessel communication.

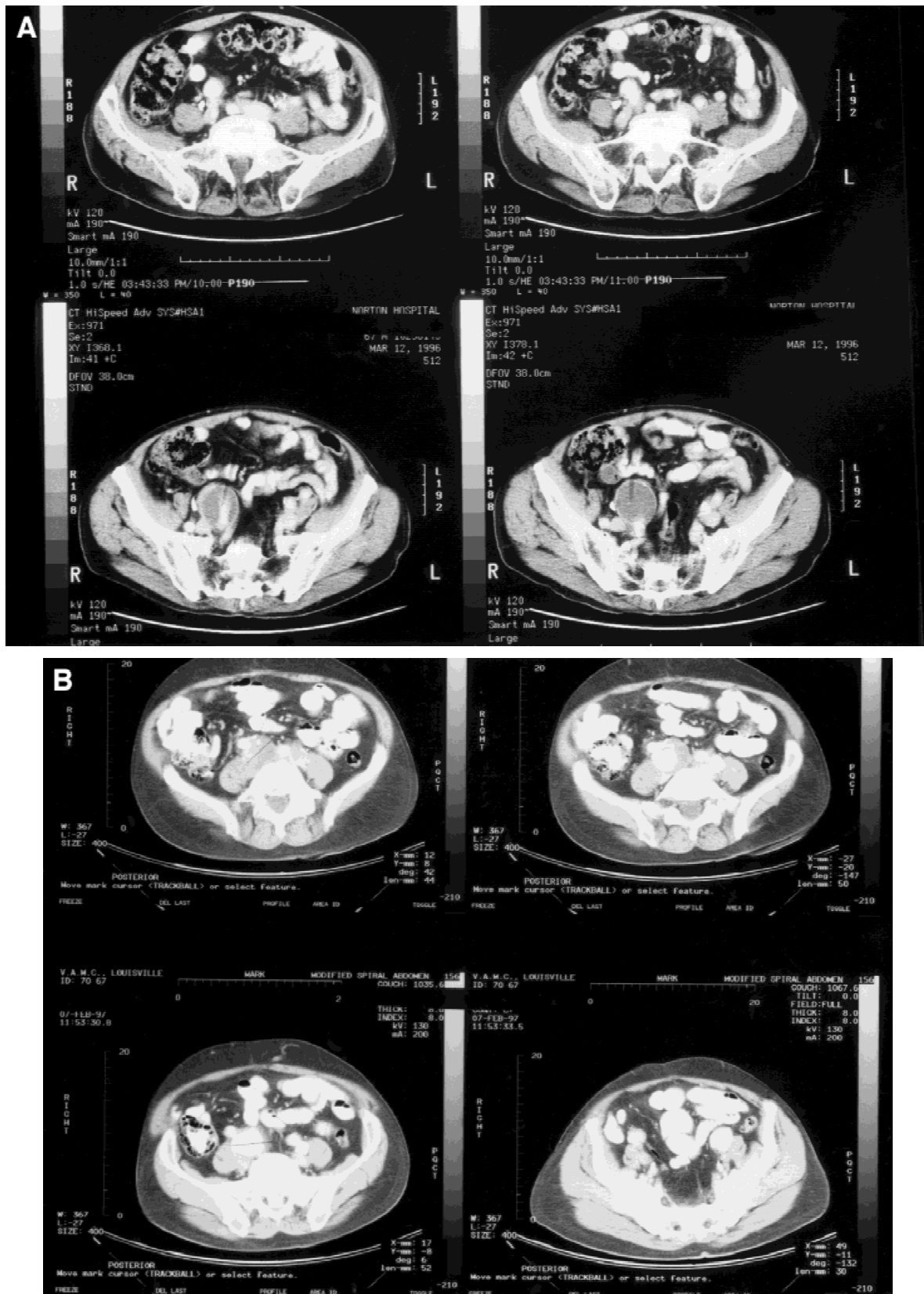


Fig. 1. Isolated common iliac artery aneurysms diagnosed by CT in an asymptomatic patient (A). Extremity swelling caused vein compression in a patient with a common iliac artery anastomotic pseudoaneurysm years after aortobi-iliac bypass repair of an abdominal aortic aneurysm (B).

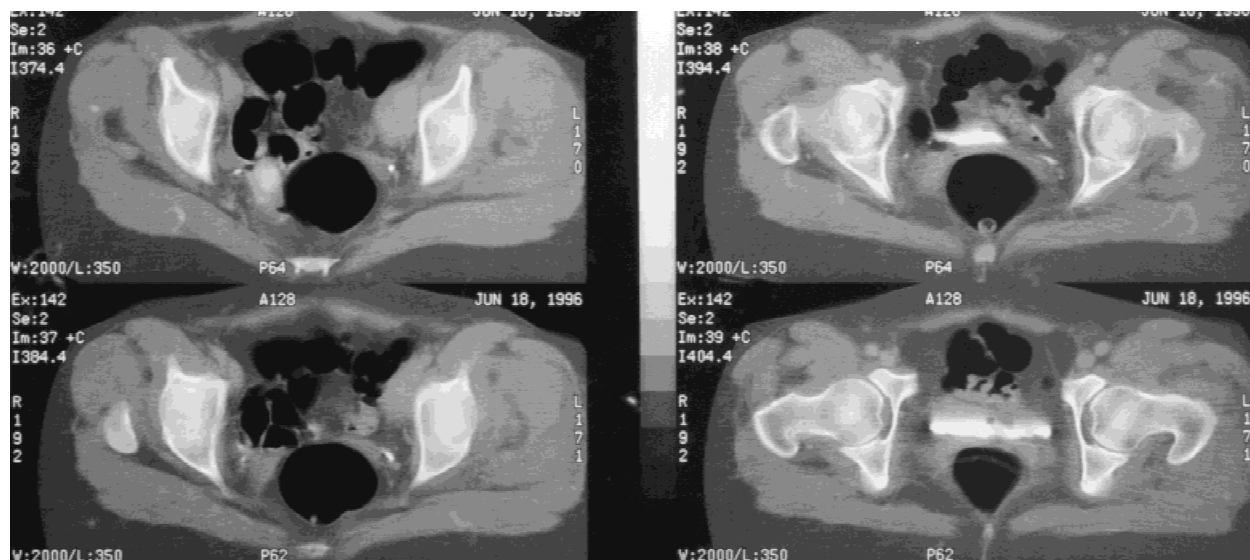


Fig. 2. A pelvic mass incidentally found by CT. An iliac vein aneurysm was confirmed by MRI and duplex ultrasonography.

They differ from hemangiomas in that they are not neoplasms. Typically, arteriovenous malformations of congenital origin are single lesions but can be multiple in certain disorders such as Rendu-Osler-Weber syndrome. The pelvis is a more common site for vascular malformations. The vascular malformations may be asymptomatic or symptomatic, most commonly appearing as pelvic or buttock pain, depending on the location of the lesion (Fig. 3A). Pelvic vascular malformations can also occur with sexual dysfunction (Fig. 3B), bleeding, leg pain, or occasionally heart failure. Malformations can be supplied by both the anterior and posterior branches of the internal iliac artery [10]. Vascular malformations in early childhood may first appear with limb growth discrepancy, the limb with the malformation being larger and longer than the opposite leg. The diagnostic test of choice is MRI with or without CT (Fig. 3C). In patients who are symptomatic and who require intervention, arteriography may be helpful.

The treatment of congenital arteriovenous malformations is extremely complex. Indications for intervention include bleeding, ischemia, intractable pain, and congestive heart failure. In children, limb length discrepancy and size of the malformation are relative indications for treatment [11]. Symptomatic lesions that are deemed resectable should be considered for complete surgical removal, because this is the only chance for cure. All other lesions should be treated by transcatheter embolization. When the patient is symptomatic, permanent embolization of the distal microvasculature of the lesion is advised. When symptoms recur, repetitive embolization is needed for treatment to control symptoms [11,12]. Occasionally, total extirpation of the congenital vascular malformation of the pelvis can be achieved with success, sometimes necessitating removal of adjacent organs

[13,14]. Ligation of the large feeding artery to the arterial venous malformation is not indicated because it prevents further embolization when recurrence and symptoms occur [15]. Intra-arterial embolization immediately prior to surgical resection is sometimes useful in controlling the blood loss during surgical removal.

### Hemangiomas

Hemangiomas are benign vascular neoplasms characterized by a large number of vessels. They are typically classified as capillary, cavernous, and deep soft tissue neoplasms. Capillary hemangiomas most commonly occur in the skin and soft tissues of the head and neck area. Cavernous hemangiomas occur most commonly in the skin and viscera. Deep soft tissue hemangiomas are the most common neoplasms of the flank and pelvic region and are classified as capillary, cavernous, and mixed patterns. The cavernous pattern predominates in most series; deep soft tissue hemangiomas may also occur within the muscle of the pelvis [16,17]. Intramuscular hemangiomas account for less than 1% of all benign vascular tumors. They have been reported in the peri-spinal muscles and gluteus maximus, with feeding arteries usually arising from the lumbar arteries. The majority (80–90%) of cases occur in patients younger than 30 years old [1,16,18]. In the pelvis, the differential diagnosis typically involves other mesenchymal neoplasms including vascular sarcomas. The patients may be asymptomatic and usually have a painless mass. Occasionally, patients with hemangioma have thrombocytopenia or Kasabach-Merritt syndrome. This has been demonstrated in intramuscular and ovarian hemangiomas. Hemangiomas and thrombocytopenia have also been reported in association with pregnancy. Preoperative diagnosis is based on





Fig. 3. Venous malformation of the pelvis diagnosed by arteriography in a patient presenting with a buttock mass (A). Vascular malformation of the pelvis in a patient with pain on intercourse and a pulsatile mass on bimanual examination of the vagina. Arteriography confirmed a congenital arterial venous malformation of the internal iliac vessels (B). MRI of a 1-year-old presenting with leg swelling with an arterial venous malformation of the iliofemoral vessels (C).

physical examination and findings on CT scan and MRI as prompted by symptoms [19]. Angiography may identify the feeding vessels to the hemangioma. The treatment of choice is total excision of the tumor [16–18,20]. A multiple modality approach with interventional radiology and surgery for embolization and surgical resection, as well as consultation with a reconstructive surgeon for closure of the defect following resection, is often the optimal treatment approach. In some cases, preoperative embolization of the feeding vessels can significantly reduce the blood loss. Upon exploration, hemangiomas have a reddish brown appearance and often have poorly defined margins. Inadequate excision is thus the most common reason for recurrence of the neoplasm. Patients with ovarian hemangioma should have total abdominal hysterectomy and bilateral salpingo-oophorectomy. If surgical resection cannot be performed because of the extensive nature of the lesion, radiation can sometimes be of use in controlling the symptoms, particularly in patients with Kasabach-Merritt syndrome and thrombocytopenia.

### Lymphocysts

Lymphocysts are benign lymphatic fluid collections that occur in the retroperitoneum and pelvis following radical pelvic surgery [21–23]. The incidence of lymphocyst formation following pelvic surgery is approximately 25%. The only factor found to be associated with the formation of a lymphocyst is the presence of a poorly differentiated neoplasm. The late occurrence of a pelvic lymphocyst should alert the treating physician to the possibility of a malignancy; surgical biopsy of the cyst wall is most useful in determining whether the lymphocyst is malignant [24]. The symptoms encountered and treatment options used depend on the size of the lymphocyst. Small lymphocysts will typically resolve spontaneously with time and require no further treatment. Larger lymphocysts can cause ureteral compression, urinary frequency, constipation, venous obstruction, and rarely precipitate deep vein thrombosis predisposing to thromboembolism [25]. Larger lymphocysts not causing significant symptoms should be followed because they will most likely spontaneously regress with time [23]. Symptomatic lymphocysts should be treated with CT- and ultrasound-guided percutaneous aspiration [26]. Repeat aspirations are frequently needed. Meticulous sterile technique should be used to prevent secondary infection of the lymphocyst. Treatment of the infected lymphocyst most frequently requires antibiotic therapy combined with surgical drainage.

### Angiomyxoma

Angiomyxoma is a benign neoplasm occurring most frequently in the pelvis and peroneal area of women, but cases in men have also been reported. It is derived from

the fibroblast cell line and consists of myxoid stroma, with mesenchymal spindle-shaped and stellate cells and prominent vascularity. Patients with angiomyxoma of the pelvis and peritoneum usually have a mass. The neoplasm can be found in multiple locations including the groin, pelvis, pubic bone, vulva, vagina, labia, ischioanal fossa, gluteus, thigh, abdomen, and cervix [27,28]. Angiomyxomas are commonly misdiagnosed and have been treated as Bartholin cysts, peroneal abscesses, and hernias [28,29]. The best method of diagnosis for angiomyxoma is a combination of CT scan, MRI, or angiography. The treatment of choice is surgical resection. Angiomyxomas are immunoreactive for actin staining and also have the presence of both estrogen and progesterone receptor proteins [29,30]. Local infiltration and ill-defined borders of the tumor are common. Recurrence of the angiomyxoma is reported in greater than 50% of the cases in the literature and emphasizes the importance of wide surgical margins of excision [28,30].

### Hamartomas

Vascular hamartomas are benign neoplasms that contain abnormal vascular channels and dilated lymphatic vessels. Patients are asymptomatic, and the hamartomas typically appear as a mass or swelling. They have been reported to occur in the pelvis and perineum as well as in the buttock region [31]. Diagnosis can be a challenge and has been confused with inguinal hernias. It is the most common soft tissue vascular neoplasm in infants and children. Juvenile hemangiomas regress spontaneously and require no treatment; however, vascular hamartomas usually persist, and the best treatment is surgical resection with adequate margins.

### Glomus Tumors

Glomus tumors are benign neoplasms that arise from smooth muscle cells of the glomus body located in the dermis of the skin [1]. They have been reported in areas in which no glomus bodies normally occur, including the cervix. Patients with glomus tumor present with painful, small, firm, and reddish-blue nodules. Glomus tumors have dilated vascular channels with glomus and smooth muscle cells. Surgical excision, the treatment of choice, is curative.

### Spindle Cell Hemangioendothelioma

Spindle cell hemangioendotheliomas typically occur in childhood or early adulthood with painless, slow-growing vascular nodules of the extremities, more commonly in the distal extremity than in the pelvic area. The tumors are well circumscribed and located in the subcutaneous tissue and have two histological patterns, with cavernous spaces alternating with cellular areas of spindle cells [32]. The treatment of choice is surgical

resection. Spindle cell endotheliomas have a greater than 50% local recurrence rate.

## **MALIGNANT TUMORS**

### **Hemangiopericytoma**

Hemangiopericytomas originate from pericytes, cells arising in small capillary vessels that underlie the basement membrane. They typically present in adulthood at 50 and 60 years of age, but also have been reported in children. Hemangiopericytomas most commonly occur in the soft tissues and muscles of the retroperitoneum and pelvis as a painless, slow-growing mass [33–35]. Hemangiopericytomas of the pelvis can occur within the uterus and retroperitoneum. Patients with uterine hemangiopericytomas compared with retroperitoneal lesions usually have good prognosis and a more benign course, typically occurring as a painless mass, with enlarging uterus or vaginal bleeding [36,37]. Patients with retroperitoneal hemangiopericytomas most commonly have a painless pelvic mass; however, some patients suffer from back and abdominal pain and rectal bleeding. Perineoplastic syndromes with breast enlargement and hypoglycemia have been reported [38]. Diagnosis is made by physical examination, CT scan, and MRI. These tumors are hypervascular, may cause hydronephrosis in patients with pelvic tumors [39], and are treated by surgical resection. Preoperative embolization of these hypervascular tumors may significantly decrease blood loss [40]. Malignant tumors most frequently metastasize to the lung and bone [34]. Malignant hemangiopericytomas have a high rate of local recurrence; radiation following resection may be beneficial [14,41].

### **Leiomyosarcoma**

Leiomyosarcomas are tumors of smooth muscle cell origin. They have been reported to arise from and impinge major vessels of the pelvis, including the inferior vena cava, iliac vein, aorta, and iliac artery. They may also erode the vessel wall and invade adjacent structures, in contrast to intimal sarcomas, which usually grow along the vessel wall. Similar to intimal sarcomas, they also grow into the lumen of the artery or vein. Eighty to 90% of reported cases of leiomyosarcomas arising from the inferior vena cava occur in women [42]. Leiomyosarcomas originating from other vessels, including neoplasms of the iliac, renal, and femoral veins, have been reported equally among men and women. The average age of onset is 50 years. The type of symptoms caused by leiomyosarcomas arising from the vena cava and the iliac veins depend on the location of the neoplasm. Suprahepatic vena cava leiomyosarcomas may occur with symptoms and signs of hepatic failure, including jaundice, ascites, and hepatomegaly. Leiomyosarcomas involving the middle portion of the inferior vena cava (adjacent to the renal veins) may cause right upper quadrant pain

mimicking cholelithiasis, renal insufficiency, and right-sided heart failure if the tumor extends to the right atrium. Leiomyosarcomas involving the lower portion of the vena cava and iliac veins usually occur with lower extremity swelling. A CT scan or MRI with inferior vena cavography are the diagnostic tests of choice. Leiomyosarcomas of the aorta and iliac arteries typically occur with claudication, or patients may have back pain [43]. Leiomyosarcomas have also occurred following aortic bypass grafting.

The treatment of leiomyosarcomas arising from the large veins and arteries is surgical resection. Local invasion and metastases are common. The overall prognosis for patients with these tumors is poor for both venous and arterial leiomyosarcomas. Chemotherapy and radiation have not been shown to be beneficial [44].

### **Angiosarcoma**

Angiosarcomas are malignant neoplasms of the endothelial cells. They commonly involve the skin and subcutaneous tissues, forming bruised, firm, and sometimes ulcerated lesions. The most common site is the head and neck region. Angiosarcoma has also been reported to involve the breast, liver, spleen, bone, retroperitoneal cavity, intraperitoneal cavity, intracranial cavity, urinary bladder, and the chronic lymphedematous extremity [1,45]. They have been reported in patients of all ages, but most commonly occur in patients who are 60–70 years old [45,46]. The treatment of choice is complete surgical resection. Radiation and chemotherapy have minimal impact on local recurrence or long-term prognosis [38]. The 2-year survival rate is about 20%. Tumors that are less than 5 cm in diameter and tumors that can be totally excised have the best prognosis [45,47]. Metastases occurs early and is reported in up to 85% of patients.

### **Lymphangiosarcoma**

Lymphangiosarcomas are angiosarcomas that occur most commonly after mastectomy and irradiation in the chronic lymphedematous upper extremity, but may arise in the pelvis of previously irradiated patients. They arise from vascular endothelial cells and have a characteristic bluish pigmented, multiple nodular appearance. There are usually several lesions noted at diagnosis. Although wide excision may be attempted, the overall prognosis is often uniformly poor. Radiation and chemotherapy have not been shown to be beneficial.

### **Intimal Sarcoma**

Intimal sarcomas involve the large vessels of the body, including the inferior vena cava and aorta. They are pleomorphic sarcomas that often resemble other neoplasms, including leiomyosarcomas, fibrous histiocytomas, rhabdomyosarcomas, osteogenic sarcomas, and chondrosarcomas [46]. Intimal sarcomas are characterized by their

growth pattern within both the lumen of the vessel and along the wall of the blood vessel. They commonly occur in patients 40–60 years of age with peripheral occlusive or embolic complications. They can also occur with aneurysmal degeneration [43,48]. The overall prognosis is poor, with a short duration of survival.

### Hemangioendothelioma

Hemangioendotheliomas are vascular neoplasms of endothelial cell origin, with four different forms classified (Table I). Hemangioendotheliomas are differentiated from hemangiomas and angiosarcomas because they have no definitive histological criterion for malignancy, rarely have large vascular channels and spaces, but commonly have plump epithelioid cells associated with a large vein [49,50].

Epithelioid hemangioendotheliomas are the most frequent and best described, commonly involving the lung and liver. They have been reported to occur in the pelvis, often involve large vessels, and are associated with leg swelling and claudication [43,51,52]. Surgical excision of the neoplasm is the treatment of choice. The local recurrence rate is approximately 15%. However, unlike most sarcomas, metastases to lymphatics occur in approximately one third of patients [49].

Retiform hemangioendothelioma is a neoplasm of the skin that can involve the trunk and extremities, usually in younger to middle-aged adults. Wide surgical excision is also the treatment of choice; however, it has a high rate of local recurrence [53]. Malignant papillary, as well as Kaposi-like hemangioendotheliomas, involve the skin and subcutaneous tissues of infants and children. Wide excision of these vascular neoplasms is the treatment of choice; the long-term prognosis is good [1].

### Intravenous Tumors

Four other tumors that may occur in the pelvis and involve the pelvic vasculature are shown in Table I. Endometrial stromal sarcoma is a tumor that arises from the endometrium of the uterus. It has been previously denoted as endolymphatic stromal myosis [54,55]. These tumors often have retroperitoneal invasion with metastases, involve regional lymphatics, and invade veins including the inferior vena cava. The symptoms of these patients sometimes include vaginal bleeding or swelling of the extremities [56]. Surgical resection with a total abdominal hysterectomy and bilateral salpingo-oophorectomy, with resection of the retroperitoneal mass, is the treatment of choice. Both local recurrence and distant metastases are common [55]. The use of progestational drugs may be beneficial.

Intravenous leiomyomatosis, a rare neoplasm originating in the uterus, invades pelvic veins, including the iliac and inferior vena cava. It is believed sometimes to represent a progression of the more common leiomyoma of

the uterus. It invades and grows into the veins of the pelvis, appearing as a pelvic mass associated with vaginal bleeding, pelvic pain, and problems related to venous obstruction such as extremity swelling. If the intravenous tumor extends to the hepatic veins, patients may develop ascites or Budd-Chiari syndrome; tumor involvement of the right atrium can cause heart failure and death. These tumors typically occur in patients who are approximately 50 years of age and is diagnosed by CT scan, MRI, and inferior vena cavography. The treatment of choice is surgical resection, with total abdominal hysterectomy and salpingo-oophorectomy [57–59].

Renal cell carcinoma can also grow into the inferior vena cava and iliac veins [60,61]. Patients may have gross hematuria, flank pain, extremity swelling, but rarely pulmonary embolus. Diagnosis is best confirmed by CT scan, MRI, and vena cavography. Echocardiography is used when tumor growth is suspected into the right atrium. Growth of the tumor into the vena cava is not a contraindication to surgical treatment. Ten percent of renal cell carcinomas will invade the vascular lumen [62,63]. Treatment includes radical nephrectomy and resection of intravascular tumor.

Myxomas and sarcomas most commonly occur in the heart, but have been reported in the inferior vena cava [64]. Intravascular tumors may present with leg swelling, upper abdominal pain, and distal embolization. Diagnosis is often by echocardiography or CT scan, and treatment is surgical excision.

### Lymphangioleiomyomatosis

Lymphangioleiomyomatosis most commonly occurs in the lungs and mediastinum. It has, however, been rarely reported in the retroperitoneal lymphatics [65]. It usually occurs in premenopausal women with pulmonary symptoms, most commonly spontaneous pneumothorax and exertional dyspnea. Lymphangioleiomyosarcoma represents a proliferation of smooth muscle cells into lymphatic vessels. These tumors often have estrogen receptors, and treatment with oophorectomy, tamoxifen, and progesterone has been attempted. Surgical resection is the treatment of choice [66].

### Kaposi Sarcoma

Kaposi sarcoma is a vascular neoplasm that has been increasing in frequency because of the AIDS epidemic. Generalized forms include cutaneous involvement; however, it may also involve lymph nodes and visceral structures. Kaposi sarcoma originates as a variety of cell types and has dilated vascular spaces. Non-AIDS-associated Kaposi sarcoma tumor is a rare entity with a more favorable prognosis. It is sometimes effectively treated with radiation therapy, with or without surgery [59,60].



## PRINCIPLES OF SURGICAL RESECTION

Vascular neoplasms within the pelvis are often difficult surgical challenges. The confined space of the pelvis, especially in men, is oftentimes obliterated by even moderate-sized tumors. Tumors often do not become symptomatic until adjacent vital structures are impinged. Thus, most tumors are apparent only after they have grown significantly. Surgical resection is thus compromised by the bony structure of the pelvis. This is confounded by the anatomic relationship of the pelvic bones to the overlying venous and arterial structures, which are many times directly impinged. A liberal use of arteriography is encouraged in developing a map for planning the resection of the more complex tumors of the pelvis. Often the CT scan or MRI will provide vital information related to the vascularity of these neoplasms. Appropriate patients may then be selected for preoperative chemoembolization. Despite adequate preoperative preparation, blood loss may be considerable; large vascular tumors of the pelvis require adequate blood banking precautions. Although it is virtually impossible to achieve complete vascular isolation, proximal and distal control of the aorto-, vena-, iliac-, and femoral-cava and vessels may minimize hemorrhage. Vital structures such as the rectum and bladder should first be dissected free from the neoplasm and appropriate vascular control maintained. The surgeon may be faced with performing a rapid operation followed by pelvic packing to minimize blood loss; a minority of tumors will only be resectable with this approach. After resection, pelvic packing with laparotomy sponges is rarely required for persistent and resistant venous bleeding.

## SUMMARY

Vascular tumors of the pelvis are a rare entity. They comprise a heterogeneous group. The proper treatment approach is usually dictated by the preoperative imaging studies, including the CT scan, MRI, and angiography. In some cases, preoperative biopsy is useful.

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